General

Title

Amyotrophic lateral sclerosis (ALS): percentage of patients diagnosed with ALS with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (e.g., noninvasive ventilation [NIV], assisted cough).

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

Measure Domain

Primary Measure Domain

Clinical Quality Measures: Process

Secondary Measure Domain

Does not apply to this measure

Brief Abstract

Description

This measure is used to assess the percentage of patients diagnosed with amyotrophic lateral sclerosis (ALS) and respiratory insufficiency with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (e.g., noninvasive ventilation [NIV], assisted cough).

Rationale

There is evidence that implementation of noninvasive ventilation (NIV) will result in improved survival. In a randomized controlled study, patients using NIV experienced a median survival benefit of 205 days (Bourke et al., 2006). NIV was initiated based on orthopnea with a maximum inspiratory pressure (MIP) less than or equal to 60 cm or symptomatic hypercapnia. No survival benefit was seen in patients with poor bulbar function.

"Early" intervention with NIV (nocturnal oximetry demonstrating greater than 15 desaturation

events/hour) resulted in 11 months longer survival compared to controls, with some beneficial effect in bulbar patients (Pinto et al., 2003). Patients who used NIV greater than 4 hours/day survived 7 months longer than patients using the device less than 4 hours/day (Kleopa et al., 1999). Forced vital capacity (FVC) declined more slowly after introducing NIV (pre -2.2%/month compared to post -1.1%/ month) (Pinto et al., 2003) and the decline was slower in those who used NIV greater than 4 hours/day (Kleopa et al., 1999). A survival benefit of 20 months was observed in NIV tolerant patients vs. 5 months in NIV intolerant patients (Aboussouan et al., 1997). NIV is probably effective in prolonging survival and in slowing the rate of FVC decline.

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for the measure:

> NIV should be considered to treatment respiratory insufficiency in amyotrophic lateral sclerosis (ALS), both to lengthen survival and to slow the rate of FVC decline (Miller et al., 2009).
> Symptoms or signs of respiratory insufficiency should initiate discussions with the patient and the caregivers about all treatment options such as NIV, tracheostomy ventilation (TV) and support in the terminal phase. Early discussions are needed to allow advance planning and directives (Andersen et al., 2005).
> NIV may be considered to enhance quality of life (QOL) in patients with ALS who have respiratory insufficiency (Miller et al., 2009).
> NIV may be considered at the earliest sign of nocturnal hypoventilation or respiratory insufficiency in order to improve compliance with NIV in patients with ALS (Miller et al., 2009).
> Use non-invasive ventilation or invasive ventilation for alleviating breathlessness. (Caution is recommended in the use of oxygen therapy to supplement ventilator support as there is no evidence that it is advantageous to motor neuron disease [MND] patients) (Heffernan et al., 2006).
> Use non-invasive intermittent positive pressure ventilators (NIPPV) or bi-level positive airway pressure ventilators to provide non-invasive ventilation, depending upon local availability and expertise, since both have been shown to improve quality of life and survival in MND/ALS patients (Heffernan et al., 2006).
> Consider non-invasive ventilation as an initial intervention in patients with, or at risk of developing, hypercapnia (Bott et al., 2009).
> Offer a trial of non-invasive ventilation if the patient's symptoms and signs and the results of the respiratory function tests indicate that the patient is likely to benefit from the treatment (Centre for Clinical Practice, 2010).
> When oxygen saturation falls below 95% the use of noninvasive ventilation and/or strategies to aid airway clearance should be considered (Bott et al., 2009).
> Oxygen therapy should be avoided or used with extreme caution due to the risk of carbon dioxide retention in patients with neuromuscular disease (Bott et al., 2009).
> The patient should be informed about the temporary nature of NIV. Care should adapt to the changing needs of patients and carers over the course of the disease (Andersen et al., 2005).
> Tracheostomy invasive ventilation (TIV) may be considered to preserve QOL in patients with ALS who want long-term ventilator support (Miller et al., 2009).
> Mechanical insufflation/exsufflation (MIE) may be considered to clear secretions in patients with ALS who have reduced peak cough flow, particularly during an acute chest infection (Miller et al., 2009).

Evidence for Rationale

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Primary Health Components

Amyotrophic lateral sclerosis (ALS); noninvasive respiratory support treatment; noninvasive ventilation (NIV); assisted cough

Denominator Description

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) and respiratory insufficiency (see the related "Denominator Inclusions/Exclusions" field)

Numerator Description

Patients with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (e.g., noninvasive ventilation [NIV], assisted cough)

Evidence Supporting the Measure

Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

Additional Information Supporting Need for the Measure

Importance of Topic

Prevalence and Incidence

Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).
Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).
Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).
ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).
In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).
Five percent to 10% of all ALS cases are inherited (NINDS, 2013).
Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).
No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).
The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated (ALS Association, 2012)
Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009).
Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995).
The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973).
The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

Mortality and Morbidity

Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., "Drug, nutritional," 2009).
Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001).
Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS

patients. Falls are an independent predictor of adverse health outcomes (Gil et al., 2008). Fall related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment (Ringholz et al., 2005).

Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival (Elamin et al., 2011), and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with motor neuron disease (MND) (Elamin et al., 2011).

Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.

Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009)

Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo (Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996).

The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); however, there are no controlled studies of benefit or harm.

Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain (Hetta & Jansson, 1997). There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.

Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m^2 (Marin et al., 2011; Lehéricey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).

ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, & Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).

End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

Office Visits and Hospital Stays

One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to

56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from $21,574 to $24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care (Dubinsky, Chen, & Lai, 2006).

Family Caregiving

Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers (Chiò et al., 2005).
Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: the perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden (Thommessen et al., 2002).

Cost

ALS is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the U.S. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g., orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS (Ginsberg & Lowe, 2002).

Disparities

All races and ethnic backgrounds are affected by ALS (NINDS, 2013).
ALS most common in individuals 40 to 60 years old, but younger and older people can develop the disease (NINDS, 2013).
Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries (ALS Association, 2012).

Opportunity for Improvement

Survival is improved with increased proportion of use of noninvasive ventilation (NIV) from 16% of ALS patients with respiratory insufficiency to 51% (Gordon et al., 2012; Chiò et al., 2012), while over 65% of ALS patients have said that they would want respiratory support indicating that noninvasive ventilation is still underutilized in ALS patients (Bradley et al., 2004; Miller et al., "Outcomes," 2009; Rabkin, Wagner, & Del Bene, 2000). Even though published guidelines to date have made a positive

impact in doubling utilization rates, underutilization of this important treatment is still evident (Bradley et al., 1999; Miller et al., "Outcomes," 2009).

Moreover, recent studies document frequent nocturnal patient-ventilator asynchrony in patients with ALS, even when using NIV prescribed as per current American Academy of Neurology (AAN) practice parameters (Atkeson et al., 2011). These findings suggest that use of NIV per these current parameters is unlikely to provide patients with ALS optimal nocturnal ventilatory support. More attention to monitoring the quality of patient sleep and oxygenation with NIV, and reducing asynchrony, is likely to improve the efficacy of this treatment (Atkeson et al., 2011).

Evidence for Additional Information Supporting Need for the Measure

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Extent of Measure Testing

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all of its measures and encourages testing of the amyotrophic lateral sclerosis (ALS) measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these

measures and to ensure that any results available from testing are used to refine the measures before implementation.

Evidence for Extent of Measure Testing

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

State of Use of the Measure

State of Use

Current routine use

Current Use

not defined yet

Application of the Measure in its Current Use

Measurement Setting

Ambulatory/Office-based Care

Home Care

Hospital Outpatient

Skilled Nursing Facilities/Nursing Homes

Professionals Involved in Delivery of Health Services

not defined yet

Least Aggregated Level of Services Delivery Addressed

Individual Clinicians or Public Health Professionals

Statement of Acceptable Minimum Sample Size

Does not apply to this measure

Target Population Age

Unspecified

Target Population Gender

Either male or female

National Strategy for Quality Improvement in Health Care

National Quality Strategy Aim

Better Care

National Quality Strategy Priority

Person- and Family-centered Care
Prevention and Treatment of Leading Causes of Mortality

Institute of Medicine (IOM) National Health Care Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Data Collection for the Measure

Case Finding Period

Unspecified

Denominator Sampling Frame

Patients associated with provider

Denominator (Index) Event or Characteristic

Clinical Condition

Denominator Time Window

not defined yet

Denominator Inclusions/Exclusions

Inclusions

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) and respiratory insufficiency

Note: Refer to the original measure documentation for International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnosis codes and Current Procedural Terminology (CPT) Evaluation and Management (E/M) service codes.

Exclusions

Documentation of a medical reason for not discussing treatment options for noninvasive respiratory support (e.g., patient is in a coma; patient has severe cognitive impairment and cannot communicate; patient is already on appropriate respiratory support)
Documentation of patient reason for not discussing treatment options for noninvasive respiratory support (e.g., patient declines to discuss treatment options)

Exclusions/Exceptions

not defined yet

Numerator Inclusions/Exclusions

Inclusions

Patients with whom the clinician discussed at least once annually treatment options for noninvasive respiratory support (e.g., noninvasive ventilation [NIV], assisted cough)

Exclusions

Unspecified

Numerator Search Strategy

Fixed time period or point in time

Data Source

Administrative clinical data

Electronic health/medical record

Paper medical record

Type of Health State

Does not apply to this measure

Instruments Used and/or Associated with the Measure

Unspecified

Computation of the Measure

Measure Specifies Disaggregation

Does not apply to this measure

Scoring

Rate/Proportion

Interpretation of Score

Desired value is a higher score

Allowance for Patient or Population Factors

not defined yet

Standard of Comparison

not defined yet

Identifying Information

Original Title

Measure #6: ALS noninvasive ventilation treatment for respiratory insufficiency discussed.

Measure Collection Name

Amyotrophic Lateral Sclerosis Performance Measurement Set

Submitter

American Academy of Neurology - Medical Specialty Society

Developer

American Academy of Neurology - Medical Specialty Society

Funding Source(s)

Unspecified

Composition of the Group that Developed the Measure

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Financial Disclosures/Other Potential Conflicts of Interest

Unspecified

Adaptation

This measure was not adapted from another source.

Date of Most Current Version in NQMC

2012 Jul

Measure Maintenance

Unspecified

Date of Next Anticipated Revision

Unspecified

Measure Status

This is the current release of the measure.

Measure Availability

Source available from the American Academy of Neurology (AAN) Web site .

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: www.aan.com .

NQMC Status

This NQMC summary was completed by ECRI Institute on March 8, 2016. The information was not verified by the measure developer.

Copyright Statement

Production

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

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